Original Article

Apraxias in Neurodegenerative Dementias

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ABSTRACT

Background: Apraxia is a state of inability to carry out a learned motor act in the absence of motor, sensory or cerebellar defect on command processed through the Praxis circuit. Breakdown in default networking is one of the early dysfunction in cortical dementias and result in perplexity, awkwardness, omission, substitution errors, toying behavior and unrecognizable gestures in response to command with voluntary reflex dissociation where, when unobserved patient will carry out reflex movements normally. Awareness into the organicity of these phenomenas will help in early diagnosis, which will help in initiating appropriate treatment and slowing down the progression of the disease. **Aims and Objectives:** The aim was to look for the various kinds of apraxias in patients with dementia using appropriate simple tests. **Patients and Methods:** Three hundred patients satisfying Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition criteria for dementia were evaluated in detail with mandatory investigations for dementia followed by testing for ideational, ideomotor, limb-kinetic, buccopharyngeal, dressing apraxia, constructional apraxia and gait apraxias in addition to recording of rare apraxias when present. **Results:** Alzheimer's disease showed maximum association with apraxias in all the phases of the disease ideational, ideomotor, dressing and constructional apraxias early and buccopharyngeal and gait apraxia late. Frontotemporal lobe dementia showed buccopharyngeal and gait apraxias late into the disease. Cortical basal ganglionic degeneration showed limb apraxias and diffuse Lewy body disease showed more agnosias and less apraxias common apraxias seen was Ideational and Ideomotor. **Conclusion:** Recognition of the apraxias help in establishing organicity, categorization, caregiver education, early strategies for treatment, avoiding anti-psychotics and introducing disease modifying pharmacotherapeutic agents and also prognosticating.

Key words: *Apraxia, degenerative dementias, organicity*

INTRODUCTION

Praxis is ability to formulate skilled movements in a nonparetic limb by planning a schema based on stored complex representations and previously learned movements and apraxia is inability to carry out a learned motor act in the absence of motor, sensory or cerebellar deficit on command. According to Geshwind, apraxia is failure to produce a correct movement in response to verbal command, imitate an action performed by the examiner, and perform the movement correctly in response to a seen object and failure to handle an object correctly. The errors may be temporal, spatial, content or lack of response (Disconnection model of Geschwind 1965). According to Liepman there is a motor engram which is the space, time plan which is conveyed by the left parietal lobe through association fibers to central regions which includes pre- and post-central gyrus, superior frontal gyrus and the underlying

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white matter. According Leiguarda and Marsden 2000, parietofrontal system encodes reaching and grasping, and frontostriatal system encodes the sequential motor events. Praxis circuit consists of Wernicke's area which analyses the verbal request and sends it to left parietal lobe, left motor cortex, anterior corpus callosum to right motor cortex[1] [Figure 1]. Lhermitte in 1942 (Psychopathologie de la vision by Jean Lhermitte1st edition-first published in 1942) postulated that both parietal lobes mediate orienting response to a sensory stimulus in space. Right parietal lobe mediates attention to stimuli from both hemisphere and left from right hemisphere only.[2,3] The principal categories of spatial disorientation are:

1. Disorders in the judgment of location or orientation of stimuli with respect to each other and to self.
2. Impairment of memory for location.
3. Topographic disorientation and the Topographic memory.
4. Route finding difficulty.
5. Constructional apraxia.
6. Spatial dyslexia and acalculia (Benton 1969).[4]

There can be impairment of localization of single stimulus, when it is called absolute localization, or it could be difficulty with perceived spatial relation between two or more stimuli called as relative localization (Peterson and Zangwill 1944). Defective appreciation of spatial relations in visual field with or without impairment of visual localization is otherwise called visual agnosia, which is due to dissociation between relative and absolute localization.[5] There can be overestimation of distance of nearby objects and underestimation of distance for far off objects. This can be assessed using Benton visual retention test, which is sensitive to subtler defects in spatial relations. It is difficult to disentangle this influence on motor performance in space, the role of praxis, memory, and intelligence. Impairment is often found in the nondominant hemisphere. Dissociation can occur between localization disorders and topographical disorders of orientation memory in which situation the simple test of spatial localization may be normal and relative tests will show an abnormality. Well-defined types of apraxia are ideational, ideomotor, limb-kinetic, dressing, and constructional. Ideational apraxia is a situation where patient knows the object including its name, but handles it as if he has never used it before due to the overall loss of the concept regarding the motor plan for use of objects. Ideomotor apraxia is a situation where patient can identify the object, knows its use, therefore, takes the right object, but does not know the motor sequence in carrying out the activity. Therefore, if patient is prompted at every step, he will complete the act. These are sub categorized into conduction apraxia when they have superior performance on verbal commands than imitation. This can be further subdivided into ability to imitate meaningful or meaning less gestures, and it can be transitive if defect is with tools and intransitive if present without tools being available. It is called dissociative if the performance is better on imitation and less on verbal command without object. It is called apperceptive if patient lacks the concept, which is equivalent to ideational. Limb-kinetic apraxia is a situation where patient is slow and clumsy in the use of a limb. These occur in lesions of the dominant parietal, premotor or sometimes parieto-occipital strip.[6] Dressing apraxia is a situation where patient has difficulty in dressing properly due to both visuospatial and sequencing errors and often seen in nondominant parietal lobe dysfunction. Constructional apraxia is inability to execute the right movement to form two- or three-dimensional pictures using single dimension lines. The other apraxias known are buccopharyngeal apraxias, apraxia of speech, gait apraxia, occulomotor apraxia, apraxia of eyelid opening, arm apraxias, etc. In patients with dementia, several kinds of apraxias are seen which is often mistaken as nonorganic in the absence of demonstrable cognitive or other deficits. As the disease advances, the diagnosis of organicity becomes easy, and most of the patients suffer severe nutritional deficiency due to buccopharyngeal apraxias and locomotion related dependence due to gait apraxia. Left arm apraxia is seen in corpus callosum lesions, which disconnects the praxicons in the left parietal lobe from the movement plans located in the right frontal lobe needed to move left hand. Right parietal lobe interferes with the sense of direction, position and movement of the left hand. Finkelinburg in 1870 described a catholic who forgot to draw a cross and carried his hand behind the ear, neck and was suspected to have a conceptual thinking problem. Steinthal 1881 described a patient who used objects as if he had never used them before.[7] Liepman postulated that this is due to inability to transform

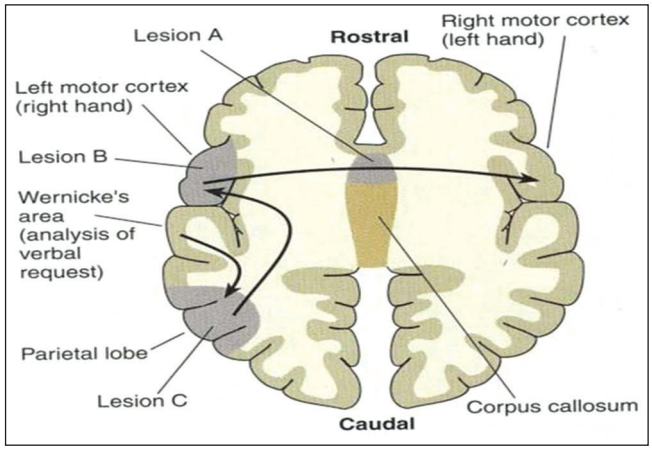

Figure 1: Leipman's praxis circuit

the image of intended actions with appropriate motor commands and neither a motor, cognitive cause could be implicated and probably represents something in between.[7-9] Deleuze 1990, postulated a person imitating the movement of another provides the model for an action automatically. Therefore, when a schema is provided externally, problem can be due to executive defect and called it as visuo-imitative apraxia. This is also seen when patients retrieve action from long-term memory, e.g., pantomimes of tool use. Rothi *et al*. 1991, postulated two pathways. A direct route from the vision to motor control accommodating both meaningful and meaningless gestures. An indirect route for familiar meaningful gestures for which ability to recognize the gesture and pull it out from the store of familiar gestures is needed. Morlass in 1928 postulated imitating meaningless gestures involves comprehension and reproduction of body centered spatial relationships.[10] Goldenberg and Strauss in 2002, said that perception and replication of meaningless gestures involves a coding of the demonstrated gestures with reference to the classification of body parts and knowledge of the boundaries defining them. Body part coding reduces the multiple visual features of the demonstrated gesture to one of a simple relationship of one body part to the other independent of the different modalities of perception. The difficulty increases with involvement of increasing number of body parts.[10,11]

Orienting apraxia is difficulty in orienting one's body with reference to objects and space (Roy and Squares cognitive model of limb praxis) Dyssynchronous apraxia indicates failure to combine simultaneous preprogrammed movements. Allokinesia is the term used to represent a situation where patient moves the body parts in the wrong direction or respond with wrong direction to commands, seen in nondominant parietal lobe. We analyzed apraxias in patients with dementia with the objective of whether it can be used as an early biomarker to categorize the dementia, the influence of intervention in reducing patient and caregiver burden and also to know if an insight can be produced among medical persons regarding the organic nature of certain bizarre symptoms in these patients.

PATIENTS AND METHODS

Patients who fulfilled Diagnostic and Statistical Manual of Mental Disorders, Fourth Edition (DSM-IV) criteria for dementia and were willing to participate in the study were taken. They were then categorized into frontotemporal lobe dementia (FTD), Alzheimer's disease (AD), diffuse Lewy body disease (DLBD) and cortical basal ganglionic degeneration (CBGD) [Figure 2a-d] using Lund Manchester criteria, NINDS-AIREN criteria for AD, consensus guidelines for the clinical and pathologic diagnosis of DLB and criteria

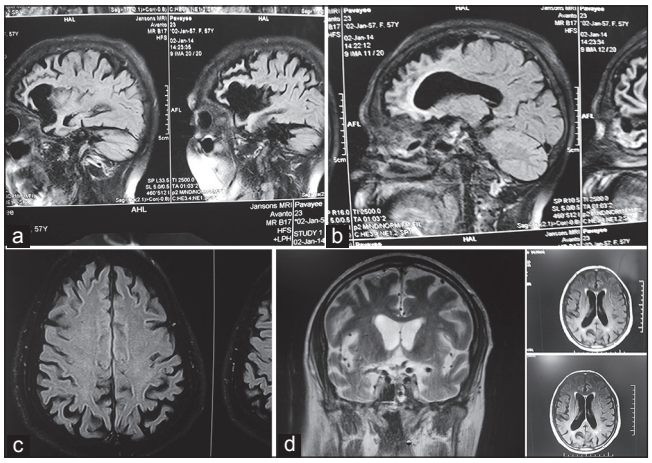

Figure 2: (a) Dragon shaped dilatation of the frontal and temporal horns in frontotemporal lobe dementia (FTD). (b) Bulbous dilatation of the frontal horn in FTD. (c) Biparietal atrophy in Alzheimer's disease. (d) Asymmetrical ventricular dilatation in cortical basal ganglionic degeneration

for diagnosis of CGBD for CBGD.[12] Apraxia was tested with verbal, visual, tactile presentation of test objects, imitation of gestures, transitive/intransitive and meaningless nonrepresentational/meaningful representational gestures, single versus multiple step tests. Common tests used for ideational was use of the comb, brush, and spoon. For ideomotor, a matchbox, candle and a board was provided and asked to light. Then construction of the clock, flower pot, crossing pentagons, and three-dimensional cube as well as wearing sari, shirt was tested. Buccopharyngeal apraxia was tested using the standard oral questionnaire which involve 20 questions (Darley 1969):

1. Open your mouth
2. Stick out your tongue
3. Blow
4. Show your teeth
5. Pucker your lips
6. Touch your nose with your tongue
7. Bite your lower lip
8. Whistle
9. Lick your lips
10. Clear your throat
11. Move tongue in and out
12. Click teeth together
13. Smile
14. Carry tongue to the top of the mouth
15. Chatter teeth like cold
16. Touch your chin with your tongue
17. Cough
18. Puff out cheek
19. Wiggle tongue side to side
20. Hum

Gait was tested using sit, stand walk assessment of the primary four parameters involving anti-gravity stretch,

stepping, propulsion and equilibrium in addition to visual assessment of stance, pace, turns, ability to climb up and down. Also asked to imitate kicking a ball and putting out cigarettes. Gait lab was not used as patients were having dementia, and it involves time in making the patient understand what is expected from them. Other Neuropsychological tests done include: (1) Motor speed (digit symbol substitution test). (2) Category fluency (animal names). (3 and 4) Working memory (verbal N-back tests). (5) Planning (Tower of London). (6) Set shifting (Wisconsin card sorting test). (7) Response inhibition (Stroop test). (8) Verbal comprehension (Token test). (9) Verbal learning and memory (auditory and visual learning test). (10) Visuo-spatial construction (complex figure test).[13,14]

RESULTS

A total of 300 patients were studied with 211 males (70.3%) and 89 females (29.67%) and the mean age of 62.5 years. Of these 68.7% were FTD, 25% with AD 3% DLBD and 3.3% with CBGD. Males constituted maximum number among FTD and AD groups, but females dominated in the DLBD group and there was no gender difference among CBGDs [Table 1]. Patients with AD showed maximum incidence of apraxia, the most common in the early phase being constructional and dressing apraxia [Video 1] and as the disease evolved developed ideational and ideomotor apraxia. Limb-kinetic apraxia is seen in only one patient and is the least common. After 3 years into the disease patients developed buccopharyngeal apraxia resulting in severe swallowing and speech difficulty and gait apraxia limiting locomotion. Patients with FTD did not suffer from apraxias in the early stage but as the disease progressed over 3-4 years, buccopharyngeal apraxia and gait apraxia became symptomatic. In patients with DLBD, early defect in construction, ideational and ideomotor apraxia is the only apraxia seen in 50% of our patients. Patients with corticobasal ganglia degeneration showed early shift of hand use in either the dominant or nondominant hand with clumsy trajectory in 70% of the patients. One patient showed the classical "pusher" phenomena characterized by drift during arm abduction with twisting of the hand at wrist resulting in diagnostic confusion as fatigability and dystonia which was excluded with negative repetitive nerve stimulation and dystonia was excluded as the tone in the limb remained unaltered. One patient had very interesting trajectory apraxia in the form of bending the whole trunk forward along with the limb when asked to bring the hand down from above head position. Then brings the trunk up keeping the limb in the down position. This patient in addition had apraxia of eyelid opening [Video 2].

Table 1: Patients with degenerative dementias over a span of 4 years

Diagnosis	Visuo spatial orientation	Ideational apraxia	Ideomotor apraxia	Limb-kinetic apraxia	Constructional apraxia	Dressing apraxia	Buccopharyngeal apraxia	Gait	Other apraxias
AD - 75 (males - 57, females - 18)	75	31	39	1	60	49	69 (after 3 years of diagnosis)	63 (after 3 years of diagnosis)	Nil
FTD - 206 (males - 147, females - 59)	Wandering without visuospatial disorientation 72 patients, utilization behaviour - 81						52 (after 3 years of diagnosis)	40 (after 3 years of diagnosis)	
DLBD - 9 (males - 2, females - 7)	4 agnosias for persons - 7	4	4		4				
Corticobasal ganglia degeneration - 10 (males - 5, females - 5)				7 (with shift of hand use)			4 (after 3 years of diagnosis)	7 (after 3 years of diagnosis)	Pusher effect - 1 Trajectory apraxia - 1

DLBD – Diffuse Lewy body disease; AD – Alzheimer's disease; FTD – Fronto-temporal dementia

DISCUSSION

This 4 years follow-up assessment of patients with dementia reveals that at least in the hospital-based population, FTD is the most common among degenerative dementias. Both in FTD and AD males dominate. FTD patients have a lower age of onset from 35 to 75 years, whereas Alzheimer's patients usually become symptomatic in the late fifth decade. The other two groups have very few numbers, but females were more in DLBD and no gender difference observed among CBGD group. Their age group varied from fifth to seventh decade. This study reveals that apraxias of all types are common in AD in early stage itself interfering with their functional efficiency. This can be explained by significant parietal lobe changes early in the disease. As the disease advances, buccopharyngeal and gait apraxias get added due to involvement of corticocortical circuits between parietal lobe and frontal lobe resulting in secondary complications. In patients with FTD, apraxias are seen only in late stage, and they are mostly buccopharyngeal and gait related which could be due to involvement of frontal subcortical circuits as the disease advances. CBGD and DLBD have variable association with apraxias, but gait and buccopharyngeal apraxias appear less common compared to the other dementias. Buccopharyngeal apraxias are disturbances of volitional movement of tongue, jaw, and lips during nonspeech tasks, whereas apraxia of speech are situations where there is impaired ability to execute voluntarily the appropriate movements of articulation of speech in the absence of other deficits. The clinical semiology is unique in these situations as they can be inconsistent with less error in involuntary speech and more errors in voluntary speech, and there can be errors of substitution, addition, deletion, distortion, repetition, and simplification. They may show effortful groping for words and put them in the wrong sequence. Imitative responses are poor, but they are aware of their errors. There can be prosodic disturbances due to self-monitoring of speech and anticipatory correction. These disturbances lead to a significant burden for patients and caregivers interfering with the minimal degree of communicative skills that may be preserved and also feeding. All higher level gait disorders are called gait apraxias and are a task specific gait disorder defined by Meyer and Barran in 1960 as a loss of ability to properly use the lower limbs in the act of walking, which cannot be accounted for by demonstrable sensory impairment or motor weakness, which can manifest as reduced velocity, short hesitant steps, pulsion abnormalities, wide base, poor balance, start hesitation, freezing and absence of rescue responses.[6] A distributed representation for praxis is described by Haaland *et al*. in 2000 and Buxbaum *et al*. 2007. According to them, left inferior parietal and dorsolateral frontal lobe is consistently involved in praxis. Posterior lesions produce Target and spatial errors, and the anterior lesions produce attention related errors. Both anterior and posterior produce internal limb configuration errors. According to Marsden and Pramstaller basal ganglia is extensively connected to superior parietal lobule, premotor area and supplementary motor area and results in subcortical apraxias. Anterior dementias affect gait by involving corticobasal ganglia thalamocortical loop and posterior dementias by corticocortical loop.[15,16] In posterior dementia patients present with difficulty in adopting the body to space and objects resulting in difficulty in sitting down with relative ease than getting up, sit on edge, stand in incorrect direction, face backwards, trying to manipulate in the air instead of orienting toward guiding objects.[17] If apraxias are recognized early quality of life can be improved by the various options available like transitive gesture training, intransitive gesture training, intransitive nonsymbolic gesture training, addition of visual cues to the environment in addition to environmental manipulation designed to reduce the risk of injury, encourage visual scanning of the environment, techniques to improve balance using vestibular stimulators, neck muscle vibrators, etc. Pharmacotherapy with levodopa, carbidopa, ropinirole, pramipexole, tizanidine, donepezil, rivastigmine, galantamine, and memantine also helps if recognized early. Buccopharyngeal apraxias can be treated by melodic intonation therapy making the patient sing an utterance, Gestures introduced into speaking facilitate deblocking and inter-systemic reorganization.[18]

CONCLUSION

Our study shows that FTD is the most common dementia in the hospital population as against the data available in the literature. This could be due to the increased caregiver burden because of the behavioral consequences of these patients. In both AD and FTD males dominated which could also be due to the probable gender bias in seeking treatment in the community. Apraxias are seen in DLBD and AD and agnosias are seen in all types of dementia in early stage. Females are more affected than males with reference to apraxias. Early Alzheimer's dementias present with dressing apraxia, ideomotor apraxia and ideational apraxia in the order of frequency and when advanced presents with buccopharyngeal and gait apraxias. The less common apraxias are trajectory apraxia and pusher phenomena seen in CBGD. Buccopharyngeal and Gait apraxias are often seen in the absence of other limb apraxias and early into the course of FTD. Agnosias and shift of the use of the hand is more common in early CBGD. Hallucinations and Agnosias are common than apraxias in DLBD. In general, in patients with

dementia, there is inadequate examination of praxis, gnosis, buccopharyngeal skills and gait. Even when complaint is there from caregivers, lack of clarity in the examiner in classification and terminologies leads to lack of attention to these symptoms. Moreover, there are no well-defined diagnostic criteria. Awareness into apraxias will avoid diagnostic confusion in these patients including being labeled as nonorganic. It will help in categorization, caregiver education, planning strategies for improving quality of life as well as prognostication and avoidance of anti-psychotics and use of disease modifying treatment, which may help to slow down the progression.

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